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CASE REPORT - NEUROSURGERY AND PREGNANCY

Decompressive craniectomy in cerebral venous sinus thrombosis during pregnancy: a case report

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Abstract

Pregnancy and puerperium are risk factors for cerebral venous sinus thrombosis (CVST), a condition which nowadays is treated non-operatively. Decompressive craniectomy is reserved only for emergency settings. We present a 22-year-old pregnant lady, who was transferred at the emergency department with a reduced level of consciousness, headache, and nuchal rigidity. Her MRI study showed CVST, causing hemorrhagic infarct and midline shift. She underwent decompressive craniectomy with partial removal of the hemorrhagic parenchyma. Remarkably, she recovered without any neurological deficits regardless of the substantial temporal lobe damage, while the thrombus nearly resolved using anticoagulation. Decompressive craniectomy can be life-saving in selected CVST patients, followed by anticoagulantion to augment the recanalization process.

Keywords Cerebral venous sinus thrombosis · Hemorrhagic infarct · Pregnancy · Decompressive craniectomy

Introduction

Pregnancy and puerperium are among the many risk factors that have been studied for cerebral venous sinus thrombosis (CVST) [4, 8]. Despite this fact, pregnant women with CVST are often misdiagnosed, a fact that complicates their further management and affects morbidity and mortality. A delay in diagnosis may be the result of many contributing reasons: rarity of the condition, delay in presentation, vagueness of symptoms which typically may be attributed to pregnancy itself, various restrictions when investigating a pregnant woman (e.g., CT scans are usually avoided), and the potentially low sensitivity of imaging studies [1]. In addition, there are many differences concerning the treatment of this entity [7], including the use of anticoagulation, intra-arterial thrombolysis, intra-arterial thrombectomy, or a combination of these methods. Decompressive craniectomy is not a first-line treatment and is mostly reserved for life-saving situations. We present what we believe is the first reported case in the literature of CVST in a first trimester pregnant lady treated with a combination of decompressive craniectomy and low molecular weight heparin (LMWH).

Case report

A 22-year-old woman in her 12th week of pregnancy presented in our emergency department with a reduced level of consciousness, headache, and nuchal rigidity. Five days before this event, she had six episodes of vomiting, for which she was hospitalized at a maternity clinic. Her clinical status improved, and she was discharged in good condition 2 days after admission. One day after that, she started developing headaches and nuchal rigidity which became worse until the point she was presented to us.

Her past medical history consisted of a possible joint hypermobility syndrome for which she was investigated when she was 16 years of age, but no definite diagnosis was made. Her brother had a spontaneous intracerebral hemorrhage at the age of 29, but the investigation was inconclusive.

Upon evaluation at the A&E, she was tachypneic, but afebrile with otherwise stable vital signs. She had a GCS of 12 (E3V4M5), anisocoria with a right pupil of 5 mm reactive to light and a mild right facial nerve palsy (House-Brackmann

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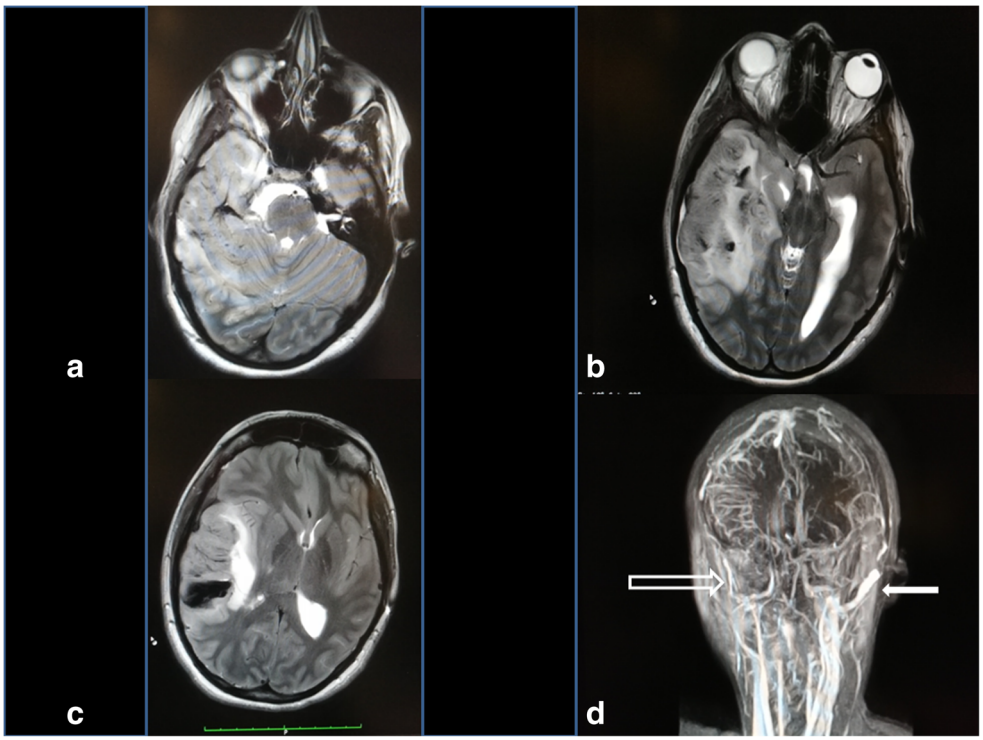

Fig. 1 **a–c** T2-weighted MRI illustrates a mixed magnetic signal lesion of the right temporal lobe causing a midline shift. **d** MRV illustrates the occlusion of the right sigmoid sinus (black arrow) in comparison with the left sigmoid sinus (white arrow)

II). She underwent an MRI scan with MRA-MRV protocol which demonstrated complete thrombosis and occlusion of the right transverse sinus to the right internal jugular vein, causing cerebral infarction with partial hemorrhagic transformation and edema of the right temporal lobe. The pathologic area was 66.7 × 45.6 × 55.5 mm (APxRLxCC) and was causing a 13-mm midline shift to the left (Fig. 1). There was no edema of the brain stem or effacement of the basal cisterns.

The patient was kept under close observation, but within 2 h, her neurological status had deteriorated to a GCS of 9 (E2V2M5). Due to the worsening of her status and the imminent coma, the patient underwent a right decompressive craniectomy including partial removal of the hemorrhagic brain parenchyma, with an effort to preserve the marginal penumbra. After surgery, she was transferred to the ICU where she was under sedation for 1 day, after which she was extubated without any major events. She exhibited a GCS of 15, stable vital signs, and spontaneous breathing with good oxygenation. Pupils returned to normal. Ultrasound monitoring of the fetus revealed no abnormalities.

The patient was then transferred to the neurosurgical ward, where she was closely monitored and investigated for the event. Anti-phospholipid antibody syndrome, inherited thrombophilia, deficiencies of anti-thrombin, protein C, protein S, or prothrombin gene mutation were excluded.

A prophylactic dose of enoxaparin was initiated 5 days after the operation and was escalated to a therapeutic body weight–adjusted dose 1 week after that.

The patient was discharged 1 month after admission with no neurological deficit (mRS[1] 0). Her mini-mental state examination was unremarkable, except for some errors at subtraction; she made no mistakes at spelling words backwards. Her follow-up MRI 2 months later showed partial recanalization of the sinuses and the jugular vein, while another MRI 4 months after the incident revealed almost complete recanalization of the sinuses and only partial obstruction of the jugular vein (Fig. 2) Remarkably, the study demonstrated substantial right temporal lobe damage, a finding somewhat inconsistent with her excellent neurological status. The patient gave birth a few days after the last MRI with a cesarean section, while the bone flap will be replaced soon.

Discussion

According to recent reviews and meta-analyses, cerebral venous sinus thrombosis is a rare, but also underdiagnosed entity, which nevertheless competes with other causes of stroke in terms of frequency during pregnancy and puerperium [10]. Pregnancy-related stroke occurs in 30.0 per 100,000 pregnancies [10]; of these, hemorrhagic and ischemic strokes occur in the 12.2 of cases respectively and CVST contributes by a rough estimate of 9.1.

Many factors have been implicated in causing or contributing to this entity; of these, prothrombotic conditions, either

[1] Modified Rankin Scale

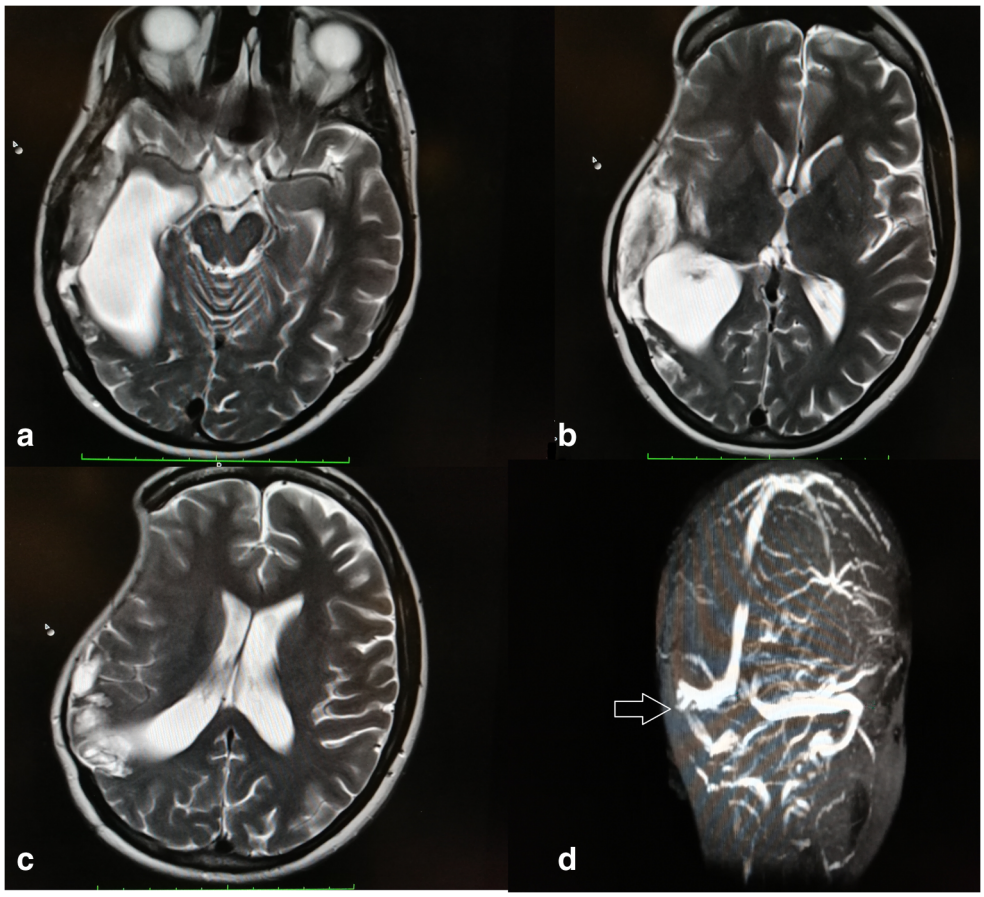

Fig. 2 **a**–**c** T2-weighted MRI illustrates post-ischemia changes of the right temporal lobe depicted as high magnetic signal lesion. Note the dilation of the right temporal horn due to porencephaly and the margins of the craniectomy in **b** and **c**. **d** TOF MRI illustrates the recanalization of transverse and sigmoid sinuses (arrow)

genetic or acquired, pregnancy and puerperium, the use of oral contraceptives, infection, and malignancy have been proposed as the most common [4, 9]. The first postpartum month in puerperium and the third trimester of pregnancy are mainly correlated to the pathogenesis of CVST, while only a few cases of CVST in early pregnancy have been reported [11].

Main determinants of good short-term outcome (mRS 0–2) differ among authors, with some defining young age [8] and others a clinical presentation with headache alone, visual field defect, and cranial nerve palsies [2]. Patients with no parenchymal lesions or hemorrhagic infarcts on radiological examination had better outcomes [2].

We presented what we believe to be the first reported case of a young lady in early pregnancy with no identifiable risk factors for CVST. In this case, the patient deteriorated quickly and immediate life-saving decisions had to be made. There are only a few cases in the modern literature reporting the use of decompressive craniectomy [3, 6, 7, 11] in pregnancy-related CVST; nevertheless, in these cases, there was a clear risk factor. Accordingly, the patient in one reported case was diagnosed with heparin-induced thrombocytopenia type II as the cause of the CVST, and she was on the last trimester of pregnancy, where CVST is not uncommon [3], while in two of the other similarly treated cases CVST took place 2 days after delivery [6] or after a missed abortion [11].

The timing and dosage of anticoagulation in the setting of hemorrhagic transformation of venous infarcts is controversial; it is further complicated in the setting of post-operative decompressive craniectomy. However, it is suggested that subcutaneous LMWH should be used in acute CVST in pregnant and puerperal patients due to its favorable safety and efficacy profile [5]; this does not apply to patients with a contraindication to LMWH or in situations where the potential of a fast reversal of the anticoagulant effect is required. The exact dosage and timing of administration have not been established in pregnant patients with hemorrhagic transformation. NOACs are not indicated for the treatment of CVST, especially during the acute phase, but this recommendation correlates with a low quality of evidence [5].

In our case, the pregnancy continued with no significant problems until delivery, but this might not always be the case; it should be kept in mind that the life of the mother is the priority during management. In addition, it is worth mentioning that a recently pooled systematic review showed that there is no beneficial effect

of therapeutic abortion [7], unless there are other medical reasons to justify it.

Concerning subsequent pregnancies, a history of CVST [1] is not a prohibiting factor. The mother should be informed that even though there is a low absolute risk of pregnancy-related venous thrombosis, the relative risk is higher compared with the general population, with 16-fold and 80-fold higher risk for VTE and CVST, respectively. There is no significant difference concerning the rate of miscarriage. In addition, prophylactic usage of subcutaneous LMWH during pregnancy and puerperium in pregnant women with a previous history of CVST is proposed [5].

In conclusion, pregnant patients with CVST usually present with non-specific signs and symptoms and sometimes there is not a clear risk factor that can be correlated to the disease. Early diagnosis is crucial to the outcome of these cases, but it is often difficult to achieve, as common diagnostic tools such as CT of the brain cannot be routinely used. Management options for these patients include the use of anticoagulation, intra-arterial thrombolysis, intra-arterial thrombectomy, or a combination of these methods. Decompressive craniectomy is recommended only when the situation becomes life-threatening by the increasing intracranial pressure with potential herniation. We propose that when performed appropriately, it can contribute to a good prognosis as the above-discussed case clearly demonstrates.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest. The patient has consented to the submission of the case report for submission to the journal.

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